Smallpox Investigation Guideline

This guideline simply outlines information about the disease management and investigation of smallpox. The user MUST refer to the state's smallpox response plan for specific information. If you suspect that you are dealing with a smallpox situation, contact the local Health Officer, on-call epidemiologist and the State Health Department immediately for assistance.

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Smallpox
Disease Management and Investigative Guidelines

CASE DEFINITION[1]

A. Clinical Description for Public Health Surveillance:

An illness with acute onset of fever ≥101º F (≥38.3 º C) followed by a rash characterized by firm, deep seated vesicles or pustules in the same stage of development without other apparent cause. Clinically consistent cases are those presentations of smallpox that do not meet this classical clinical case definition: a) hemorrhagic type, b) flat type, and c) *variola sine eruptione*. (Detailed clinical description is available on the CDC web site, see URL: http://www.bt.cdc.gov/agent/smallpox/index.asp).

B. Laboratory Criteria for Diagnosis:[2]

- Polymerase chain reaction (PCR) identification of variola DNA in a clinical specimen, or
- Isolation of smallpox (variola) virus from a clinical specimen (Level D laboratory only; confirmed by variola PCR).

C. Case Classification:

- Confirmed: Case of smallpox that is laboratory confirmed, or a case that meets the clinical case definition that is epidemiologically linked to a laboratory confirmed case.
- Probable: A case that meets the clinical case definition, or a clinically consistent case that does not meet the clinical case definition and has an epidemiological link to a confirmed case of smallpox.
- Suspect: A case with a generalized, acute vesicular or pustular rash illness with fever preceding development of rash by 1-4 days.
- Exclusion Criteria: A case may be excluded as a suspect or probable smallpox case if an alternative diagnosis fully explains the illness or

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[1] The smallpox case definition is to be used only during post-event surveillance. The case definition described in Guide A of the Smallpox Response Plan and Guidelines (Version 3) on the CDC bioterrorism preparedness website (URL: http://www.bt.cdc.gov/agent/smallpox/response-plan/index.asp) includes different criteria for a suspected case than the smallpox case definition the Council of State and Territorial Epidemiologists approved for use in the National Notifiable Diseases Surveillance System (NNDSS). The smallpox case definition on the CDC bioterrorism web site is more sensitive and less specific than the case definition for the NNDSS, in that a "suspect" case is defined as: "a case with febrile rash illness with fever preceding the development of rash by 1-4 days."

[2] Indications for laboratory testing of patients with suspected smallpox should be followed as described in detail in Guide A of the CDC Smallpox Response Plan. Laboratory diagnostic testing for variola virus should be conducted in Level C or D laboratories only.

appropriate clinical specimens are negative for laboratory criteria for smallpox.

D. Laboratory Tests:

- Only vaccinated individuals who wear gloves and masks should collect laboratory specimens from suspect cases.
- Specimens should be forwarded to a Bio-Safety Level C or D rated laboratory. The Kansas Health and Environmental Laboratory not Level D rated.
- Remarks: For additional information and/or questions concerning isolate collection, sample transport and laboratory kits call (785) 296-1620. An online manual of laboratory tests is available at http://www.kdhe.state.ks.us/labs/links.html

E. Bioterrorism Potential:

Smallpox is a potential bioterrorism weapon. If you suspect that you are dealing with a smallpox contact the local Health Officer, on-call epidemiologist and the State Health Department immediately.

F. Outbreak Definition:

A single suspect or confirmed case is considered to be an outbreak and is considered to be an international public health emergency. Because the disease has been eradicated, a single case is also considered to be a bioterrorist event until proven otherwise.

INVESTIGATOR RESPONSIBILITIES

A. Investigation Tasks and Activities:[3]

- Conduct an epidemiological investigation to identify the possible source of infection and to locate additional cases and/or contacts in the community.
- Identify the source of infection and prevent further transmission.
- Identify contacts that may have been exposed to the index case and refer them for proper prophylaxis therapy.
- Report all confirmed, probable and suspect cases to the Bureau of Epidemiology & Disease Prevention, using established methods.
- Establish and maintain a detailed line listing of all cases and contacts with accurate identifying and locating information.

[3] All epidemiological activities MUST be coordinated with state and Federal health and law enforcement agencies. Refer to the state's smallpox response plan for specific information.

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B. Notifications:

- Report by telephone all suspect and/or confirmed cases to the Local Health Officer, the on-call epidemiologist (local) and KDHE (1-877-427-7317) immediately.
- Mail or deliver notification letter and/or disease fact sheet to case, contacts and other appropriate individuals or groups (if appropriate and/or requested).

EPIDEMIOLOGY

In 1980, the World Health Organization declared that smallpox had been eradicated. The last naturally occurring case occurred in Somalia in 1977 and 2 cases attributed to laboratory exposure in 1978. The United States discontinued routine childhood immunization against smallpox in 1971 and the immunization of health care workers in 1976. The United States military continued to immunize their personnel until 1990. Officially, smallpox is now only found in designated research laboratories sites in the United States and Russia.

DISEASE OVERVIEW

A. Agent:

Variola virus, a species of Orthopoxvirus. The agent is the same for variola major (classic smallpox) and variola minor (a less serious form of the disease).

B. Clinical Description:

The rash of smallpox is preceded by a prodromal symptoms including: a high fever lasting 3-4 days, headache, malaise, muscle pain, prostration, and occasionally nausea, vomiting, and backache. The trademark of the classic smallpox is a generalized vesiculopustular rash with lesions found more densely on the face and extremities (centrifugal), including the palms and soles. All lesions on any one part of the body usually appear as a single crop with all lesions progressing from the macular to the pustular stage at about the same time. The rash progresses from sparse macules (day 1), to papules (days 2), vesicles (days 3-4), pustules (days 5-12), and scabs (days 13-18) for a total duration of 2-3 weeks. Less common presentations of the smallpox rash include flat, or hemorrhagic lesions. A rash that progresses through the stages more rapidly and has fewer lesions characterizes modified smallpox, which occurs more commonly among previously vaccinated persons.

- **Differential Diagnosis:** Varicella, disseminated molluscum contagiosum, secondary syphilis, meningococcal sepsis and monkeypox.

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C. Reservoirs:
Humans are the only known hosts; there are no known animal reservoirs and/or vectors. Officially, smallpox is now only found in designated freezers in the United States and Russia.

D. Mode(s) of Transmission:
Highly transmissible person-to-person by inhalation of virus-containing airborne droplets of saliva or contact with material from smallpox lesions from an infected person. Transmission may occur in a hospital setting if isolation of the case is not implemented immediately. Attack rates may be as high as 50% in susceptible populations.

E. Incubation Period:
Range, 7-17 days; average, 10-14 days. Skin eruption appears 2-4 days after first symptoms.

F. Period of Communicability:
Smallpox cases are generally not infectious to others until the onset of rash (approximately 7-17 days after exposure). Since the exact date of rash onset may not be reported accurately cases should be considered potentially infectious from date of onset of fever. The period of highest transmission is during the first 7-10 days after onset of rash, however, a person is considered infectious until all scabs have separated.

G. Susceptibility and Resistance:
Susceptibility is universal among the unvaccinated. Adults vaccinated as children and military personnel vaccinated in the late 1980's are also considered susceptible. Only individuals who have been recently vaccinated or who have recovered from recent infection should be considered to be resistant.

H. Treatment:
Supportive only.

STANDARD CASE INVESTIGATION AND CONTROL METHODS
Standard investigation activities include the following: 1) Confirmation of the diagnoses (*i.e.,* case definition), 2) Collection of relevant demographic and clinical data (*e.g.*, age, sex, disease syndromes and/or symptoms), 3) Determination of the setting (*e.g.*, community, hospital, daycare or other facility), 4) Investigation of possible epidemiologic links among cases (*e.g.,* cluster, family, co-workers) and 5) Exposure histories to help prioritize delivery of vaccine and/or vaccinia IG. Most of the information can be obtained from the case person, healthcare provider and/or the medical record.

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A. Identify Potential Source of Infection:[4]

To help identify the source of the infection, the investigator should focus their investigation within the incubation period of smallpox (*i.e.,* 7-17 days).

B. Identify Potential Exposed Individuals / Populations (Contacts):[5]

Interview case (if possible) to obtain detailed name and contact information for all persons with whom the case had face-to-face contact (≤ 6 feet) since onset of fever until time of interview; include, places visited by the case since onset of fever including health care provider offices, clinics, and emergency departments; work, school, and regular, as well as occasional activities. If case is unable to answer questions because of age or illness, obtain information from case's close family members and friends.

C. Isolation, Work and Daycare Restrictions:

- Case: Place case immediately into airborne, contact isolation and standard isolation precautions; maintain isolation for the duration of disease until all scabs have separated from skin lesions.
- Contacts: Contacts are to remain under active surveillance for 18 days (one day after the longest incubation period for smallpox) after their last contact with the smallpox patient, or 14 days after successful vaccination. Contact-tracing personnel should establish methods for daily reporting by the contact. Asymptomatic, vaccinated contacts may continue routine daily activities, but should remain relatively close to of home, unless otherwise authorized in writing by the health officer or the health officer's representative, and should check their body temperatures twice daily. If resources permit, closer monitoring (*e.g.,* daily visits) by public health personnel should be considered.

D. Follow-up of Cases:

Cases should be monitored to assure compliance with isolation guidelines until they are no longer infectious.

E. Protection of Contacts:[6]

- List and prioritize all contacts for need of vaccination based on type and duration of contact as well as prior immunization history for smallpox.
- Make arrangements for the immediate vaccination of asymptomatic contacts and contact's household. If they cannot be vaccinated due to

[4] Personnel designated for case interviews and contact investigation must be effectively vaccinated prior to initiating face-to-face interviews with the case and/or contacts,
[5] Personnel designated for case interviews and contact investigation must be effectively vaccinated prior to initiating face-to-face interviews with the case and/or contacts.
[6] The prioritization and delivery of smallpox vaccine and vaccinia IG is beyond the scope of this document. The user should refer to the state smallpox response plan for specific information.

contraindications or other reasons, insure that they avoid exposure to the contact until the end of the contact's surveillance period.
- If contact is symptomatic with fever or rash, make immediate arrangements for the safe transportation of contact to the County's designated facility for evaluation of smallpox cases.

F. Environmental Measures:
None.

G. Education:
- Emphasize the importance of immunizing all contacts unless it is contraindicated.
- Instruct all cases and contacts on methods of disease transmission and communicability and the actions required to prevent further transmission.

MANAGING SPECIAL SITUATIONS
Bioterrorism:
A single confirmed case or suspect case of smallpox is an outbreak and is considered to be a bioterrorist event until proven otherwise. Contact one the following numbers immediately in order of priority as shown:
- Kansas On-call Epidemiologist 877-427-7317
- CDC Bioterrorism response coordinator 404-639-0385
- An announced threat of dissemination, though most likely a hoax, should be taken seriously and the State Health Department and the local FBI Duty Officer notified. 816-512-8200

A. Likely Bioterrorism Scenario:
If smallpox virus were to be used for a bioterrorist attack, it would most likely be disseminated in aerosol form. It may be released upon a large number of people at a gathering; however, it would also be effective if released upon smaller numbers of people in a single or multiple locations. Such an attack may or may not be announced. It is very possible that public health and law enforcement authorities would only learn about such an attack upon diagnosis of the first case.

B. Safety Considerations for Public Health and Other Health Care Professionals:
- From the moment that smallpox is suspected all health care professionals involved in the care of the patient should observe strict respiratory and contact isolation precautions. The patient should be immediately placed into strict respiratory and contact isolation. All health care employees exposed to the patient prior to recognition of the situation should receive vaccine and vaccinia IG as soon as possible and should be treated as

contacts. Subsequently, all health care workers should be vaccinated prior to caring for any cases or suspect cases.

- Only vaccinated individuals who wear gloves and masks should collect laboratory specimens of suspect cases. Specimens should be forwarded to a Bio-Safety Level D rated laboratory. The Kansas Health and Environmental Laboratory is not Level D rated.

C. Definition of the Population-at-Risk:

Defining the population-at-risk is essential to guide response activities. Public health authorities will play the lead role in this effort, but must consult with law enforcement, emergency response and other professionals in the process. The definition of the population-at-risk may have to be re-evaluated and redefined at various steps in the investigation, assessment and response to a bioterrorist event. Once the mechanism and scope of delivery has been defined, the identification of the symptomatic and asymptomatic exposed individuals can be completed and recommendations for the treatment and/or chemoprophylaxis made.

D. Specific Control Measures Include:

- Decontamination: Not necessary.
- Post-exposure prophylaxis (PEP): Recommended for all contacts.
- Isolation: Mandatory for cases for 18 days (one day after the longest incubation period for smallpox).
- Quarantine: Mandatory for contacts for 21 days, regardless of receipt of PEP.
- Line lists: A central responsibility of the investigative staff is to maintain detailed line lists of cases, suspect cases, exposed, and potentially exposed individuals with accurate identifying and locating information as well as appropriate epidemiological information. These lists will be essential for early identification of infection among the exposed.
- Pharmaceuticals: In the event of an outbreak of smallpox, vaccine and vaccinia IG will be procured from the Strategic National Stockpile. Procurement, storage, and distribution will be coordinated through the Kansas Department of Health and Environment. Local and state public health officials must play a central role in determining which individuals should have priority for receipt of limited pharmaceuticals.

ADDITIONAL INFORMATION / REFERENCES

- American Academy of Pediatrics. 2003 *Red Book: Report of the Committee on Infectious Disease, 26th Edition.* Illinois, Academy of Pediatrics, 2003.
- Heymann. D., ed., *Control of Communicable Diseases Manual, 18th Edition.* Washington, DC, American Public Health Association, 2004.

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- Case definitions for Infectious Conditions Under Public Health Surveillance, Division of Public Health Surveillance and Informatics, Nationally Notifiable Infectious Diseases, United States 2005. Available at: http://www.cdc.gov/epo/dphsi/PHS/infdis2005.htm
- Kansas Department of Health and Environment, Bureau of Epidemiology. *Disease Protocols,* 2001.
- County of Los Angeles, Department of Health, Public Health Programs and Services, *Communicable Diseases Manual*, June 2003.
- Oklahoma State Department of Health, Communicable Diseases Division. *The Epidemiologic Follow-up of Communicable Diseases in Oklahoma,* 2001.
- Missouri Department of Health and Senior Services, Section of Communicable Disease Control & Veterinary Public Health, *Communicable Disease Investigation Reference Manual*. 2001.
- Oregon Health Services Website. Available at http://www.ohd.hr.state.or.us
- Commonwealth of Massachusetts, Department of Public Health Website. Available at http://www.state.ma.us/dph/
- CDC Website. Available at http://www.cdc.gov/health/default.htm
- Smallpox Response Plan and Guidelines (Version 3.0) available at http://www.bt.cdc.gov/agent/smallpox/response-plan/#guidea

Public Health Fact Sheet
Smallpox

What is smallpox?
Smallpox is a viral disease that causes a severe illness characterized by a high fever and a severe rash. It is very contagious and up to 1/3 those who are infected may die. The last naturally occurring case occurred in Africa in 1977. Before a vaccination and eradication campaign finally brought an end to smallpox, the disease was found worldwide.

What are the symptoms?
Initial symptoms of smallpox included high fever, fatigue, headache and backache. These symptoms usually develop 10-14 days after a person has been infected. A rash usually appears 2-3 days after the fever and appears first on the face, mouth, hands, forearms, palms, or soles of the feet. It then spreads to the legs and the trunk and progresses to raised bumps and pus-filled blisters. These blisters crust, scab, and fall off after about 3 weeks, often leaving a pitted scar.

How is smallpox spread?
Smallpox is spread person-to-person in two ways. A person can inhale particles containing the virus that have been coughed into the air by an infected person, or a person can be infected when touching the rash or blisters of an infected person and then touching their own nose or mouth. Contact with the clothing and bedding of an infected patient can also spread the virus.

Who gets smallpox?
Susceptibility to smallpox is universal among the unvaccinated. Adults vaccinated as children and military personnel vaccinated in the late 1980's are also considered susceptible. Only individuals who have been recently vaccinated should be considered to be resistant.

How is it diagnosed?
Smallpox can be diagnosed based on a patient's clinical signs and symptoms; laboratory tests are also used. Laboratory testing needs to be completed in specialized laboratories with appropriate testing techniques and measures to protect the laboratory workers.

How is smallpox treated?
There is no proven treatment for smallpox, but research to evaluate new antiviral agents is ongoing. Patients with smallpox may receive supportive therapy such as intravenous fluids, medicine to control fever or pain and antibiotics for any secondary bacterial infections that may occur.

How can smallpox be prevented?
There is a vaccine to prevent smallpox that was routinely administered in the United States until the early 1970's. The risk of adverse events resulting from the vaccine, accompanied by the rapid decrease in smallpox around the world was part of the justification to discontinue routine vaccination against smallpox before the disease was eradicated in 1977. Vaccination of the general public is not recommended at this time.

If someone is exposed to smallpox, is it too late to get a vaccination?
No, vaccination within 3 days after exposure will probably completely prevent or prevent the disease from becoming more severe in the vast majority of persons. Vaccination 4-7 days after exposure likely offers some protection from disease or may modify the severity of disease.

If I received a smallpox vaccination prior to 1972 am I still immune?
It is unlikely that it would prevent you from getting infected, but it would reduce your chance of dying. The long-term effectiveness of the smallpox vaccine is uncertain and it is generally assumed that most of the United States population is at risk for smallpox. A report from Europe suggests that people vaccinated 10-20 years ago have enough immunity to lessen their chance of death if infected. However, these people need another dose of smallpox vaccine to restore their immunity.

Where can I get more information?
- Your Local Health Department
- Kansas Department of Health and Environment, Epidemiologic Services Section (877) 427-7317
- http://www.cdc.gov/health/default.htm
- Your doctor, nurse, or local health center

This fact sheet is for information only and is not intended for self-diagnosis or as a substitute for consultation. If you have any questions about the disease described above or think that you may have an infection, consult with your healthcare provider. This fact sheet is based on the Centers for Disease Control and Prevention's Health and Safety topic fact sheets.